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Optic neuritis

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A CASE OF DOUBLE OPTIC NEURITIS

WITHOUT

CEREBRAL TUMOUR.

A CASE OF DOUBLE OPTIC NEURITIS WITHOUT
CEREBRAL TUMOUR.

By J. HUGHLINGS JACKSON, M.D., F.R.C.P.

In the case I have to relate there was well marked double optic neuritis with blindness. At the autopsy no morbid intra-cranial changes were seen except those of very great congestion of the brain, and these were doubtless recent. In this regard the case is exceptional. In most cases of double optic neuritis there is tumour, or some kind of "coarse"* disease. In the case I am about to relate there were (see Dr. Sutton's examination, p. 10) minute morbid changes; but still the case is very exceptional.

It must be remarked too, that although I speak chiefly of the optic neuritis, *the whole tenor of the case was that of a case of intra-cranial tumour.*

It would, I believe, have been considered a case of cerebral tumour by physicians who do not value ophthalmoscopic evidence.

I have now seen several cases of double optic neuritis in cases without cerebral tumour.

Nevertheless I think that optic neuritis *is the most valuable of all signs of coarse organic disease within the cranium*; but it is not decisive evidence. The following quotation from a "Lecture on Optic Neuritis," published in

* The expression "coarse disease" is used as a general term to include new growths (gliomata, syphilomata, &c.) cysts, abscess, and in fact all kinds of adventitious product, or to use a common word, all sorts of lumps within the cranium. These are all "foreign bodies," and each of them may lead to changes in the optic nerves.

I think too that there is but *one* kind of change produced by intra-cranial growths; I call this optic neuritis. I do not make the distinction into optic neuritis and "stauungs papilla" ("choked disc" of Allbutt.)

1871, embodies the opinions I still hold on the diagnostic value of neuritis. I begin by speaking of the limitation of my experience of optic neuritis.*

"I have spoken above of a Physician's experience, and I hasten to qualify my remarks. I see those patients who have *severe cerebral disease*. No one ever consults me for defect of sight only, but for such symptoms as severe headache, convulsion, and hemiplegia, with which optic neuritis often occurs. To me defect of sight is but one symptom, and not the most important one, in a series. I admit, then, that my experience is of necessity one-sided. Further I admit that in a few cases of severe cerebral disease where I have discovered double optic neuritis, I have found no kind of coarse disease *post-mortem*.

"I have been wrong several times in the diagnosis of an adventitious product within the skull in cases where there had been found double optic neuritis, *but I have far oftener been wrong by neglecting the inferences above stated to be deducible from the presence or absence of optic neuritis*—wrong in saying there *was* an adventitious product when the discs were normal, and wrong in saying there *was not* when there was double optic neuritis. I feel, therefore, justified in saying that double optic neuritis does point *very strongly indeed* to coarse disease inside the head.

"You will not misunderstand me to imply that you are to diagnose tumour or other coarse disease of the brain solely by the ophthalmoscope. You have, in most cases, *no need* to rely on this one condition. You do not diagnose phthisis by the physical signs alone. You may say that you would be right *in most cases* if you did trust to physical signs alone. I may go so far as to the ophthalmoscopical signs: You would be right *in most cases*, I believe, if you diagnosed coarse disease within the skull by the presence of optic neuritis."

* This and the five subsequent paragraphs have already been published in a pamphlet printed for private circulation. They appeared in this way before the case related in the text was seen by me.

In some cases we find with optic neuritis (I now speak of cases in which *post-mortem* examination reveals intra-cranial tumour) changes in the retina very like, sometimes quite like, those occurring with chronic Bright's disease. On the other hand, in some cases of Bright's disease the disc is swollen to a degree equalling that found in some cases of intra-cranial tumour, and there are lustreless and shapeless white spots about the disc, and also blotches; not only streaks of blood like those which are seen in very many cases of neuritis from intra-cranial tumour. Dr. Herm. Schmidt and Dr. Wegner ("Archiv. f. Oph.," Bd. xv, Abth. iii, s. 253—275) report a case of cerebral tumour in which the ophthalmoscopic signs were quite like those in a case of kidney disease. (See Dr. Noyes' report on "Ophthalmology," "New York Medical Journal," February, 1871, p. 210.)

Hutchinson ("Royal London Ophthalmic Hospital Reports," vol. v, p. 308), in some cases of optic atrophy in children, no doubt the sequel of neuritis, has seen at the yellow-spot region groups of highly refractive globules, resembling at first glance clusters of spider's eggs.

The following is an extract from Mr. Brudenel Carter's recently published work. I fear I blundered in not having examined the patient's urine in the case of renal disease to which he refers.

"Many attempts have been made by various writers to distinguish 'neuritis optici' from 'perineuritis,' and both from mechanical obstruction; but I have never been able to satisfy myself of the validity of the distinctions which have been drawn. Dr. Hughlings Jackson, who has investigated the subject with infinite pains and care, and who has followed a large number of his cases to the *post-mortem* table, will not say more than that 'optic neuritis,' his general expression for the changes under consideration, is an evidence of some coarse intra-cranial lesion, and I myself can only say that it is frequently associated with such lesion.

"We had a little boy in St. George's Hospital in 1872,

who was transferred from my care to that of Dr. Fuller, and who had choked discs of the most typical character. Dr. Hughlings Jackson saw him, and entertained no doubt that he was the subject of some form of brain disease: and the same opinion was expressed by Dr. Noyes, of New York, and by several members of the International Ophthalmological Congress, which was then assembled in London. The boy died of pleurisy supervening upon advanced kidney disease, and no trace of mischief in his brain could be discovered by the most careful examination.

“Nearly at the same time we had in the Hospital a young woman whose eyes presented typical examples of the changes often associated with albuminuria, but who died with healthy kidneys, of a tumour in the cerebellum.”

One peculiarity in the case I am about to relate is the absolute amaurosis. This is a rare thing in physicians' practice. I rarely see such discs as this patient had, with loss, or even with extreme defect, of sight. On the other hand I very often see optic neuritis without impairment of vision. The patient was sent to me by Mr. James Adams, it was then essentially an ophthalmic surgeon's case.

I would say, too, that during the woman's life I had no doubt that we should find an intra-cranial tumour (or other kind of coarse disease). I ought to have been less confident, because I once before felt certain that I should find an intra-cranial tumour in a case of double optic neuritis, and found only general cerebral atrophy; there was no adventitious product, nor any trace nor relics of it. I did not examine any of the brain of that patient microscopically.

In the case about to be narrated we cannot invoke raised intra-cranial pressure as a cause, for there was no evidence of it; the congestion of the brain was certainly no evidence of it; it was simply owing to the mode of death. I cannot do better here than quote what Wilks and Moxon say of cerebral congestion.

“There is no doubt that congestion of the brain is the cause of serious symptoms, and even of death. We particularly ask

you not to suppose that we are denying the existence of congestion of the brain as a fatal disease, what we wish to say is, that a fulness of the vessels after death cannot *prove* the occurrence of the primary congestion, and in fact *post mortem fulness of the vessels of the brain and its membranes is of no pathological value whatever* (italics in original) as showing brain disease, because asphyxia produces it. On the other hand anæmia of the brain does exclude congestion, and also proves that death was not through asphyxia, which always produces congestion. Our reason for saying all this so confidently, when even now you find writers in systems of medicine speaking of redness of the cerebral membranes as the great sign of inflammation, &c., is that we have seen all degrees of intensity of such congestion when there was no trace of cerebral disturbance during the life of the individual, and this we have seen, not occasionally, but as a common occurrence. Again, it is not only the experience of morbid anatomists which declares intra-cranial congestion to be thus valueless, we will quote for you a passage from Kussmaul and Tenner's account of their elaborate researches on the effects of hæmorrhage as causing epilepsy. They say, 'We could never deduce any results from the *post mortem* examinations undertaken with a view to determine the state of fulness before death of the most important parts of the vascular system, viz., of the arteries and arterial capillaries, and even in the most favourable instance, when similar inquiries were directed towards the veins, our results could only be looked upon as approximate.'

"Thus you will see that it is only for lack of information that any one would speak of congestion of the brain as a proof of a cause of death. You may *infer* reasonably from the circumstances of particular cases that death occurred from congestion of the brain, but you cannot *prove* it after death by inspection. We shall presently endeavour to show you in what degree certain appearances of the brain are evidences of congestion." ("Pathological Anatomy," Wilks and Moxon, pp. 202-3.)

SUMMARY OF CASE.—*Headaches nearly all life—Attack of Vertigo with Unconsciousness—Intense Headache, Vomiting and Double Optic Neuritis—No Paralytic Symptoms—Death in Asphyxia—Autopsy—No “Coarse” Disease—Extreme Cerebral Congestion.*

(Abstract of Notes by Mr. Charles Mercier.)

Henrietta S., æt. 34, admitted December 19, 1874, married. Her family history presented nothing of importance.

Ever since she could remember she had suffered with pain in her head, which she described as of two kinds—headache, and peculiar pain, darting like neuralgia or “rheumatics.” The former occurred generally about once a-week, the latter usually after her menstrual periods. No history of syphilis.

Three years before admission her husband came home from a long absence at sea, and three months later she had a miscarriage. From this time her menstrual flow became exceedingly irregular, coming on at the slightest provocation at any time, and lasting for weeks together. A year and three quarters later she had another miscarriage, and a year before admission her head began to be very bad. The first occurrence of her illness was an attack of severe vertigo, with momentary unconsciousness, followed by severe headache which lasted for four days. At the end of this time she had another attack, after which the headache lasted for three weeks, with occasional remissions, during which she vomited. The vomiting was usually followed by a feeling “as if she were going down into a pit,” and by hot sweats.

At this time also she used to have “dark shadows coming over her eyes, and popping noises all about her head, and such dreadful whistling and howling in her ears.”

After three months the condition of her sight was as follows. Sometimes she had an appearance of darkness, and at such times, although her sight was very deficient, she could, although with difficulty, recognise people’s faces. At other times she had an appearance of light, with heavy shadows waving in front of her, and although, from the sensation being one of light, she thought that her sight was better at these times, it is probable that vision was quite absent, for she used to run against objects. At length, three months before admission, she rather suddenly had an impression of brilliant light which remained permanent, and since that hour she was completely blind.

On admission, she was a well nourished woman, of exceedingly dark complexion; very intelligent, giving her answers with unusual precision and directness for a hospital patient, and she showed great dexterity in making up for her loss of sight by employing her ears, &c. She was by no means an emotional woman, was very patient under her suffering, which was evidently great, explained and described naturally the subjective sensations which affected her without any exaggeration of expression or gesture. Her organs and functions were all in good order, her vital condition good. Pulse regular, but very small, hard and singularly incompressible. There was no local paralysis whatever, no defect of voice, articulation or speech. She suffered occasionally from attacks of most severe pain in the head, frequently accompanied by vomiting. There was extreme double optic neuritis. Mr. Couper (January 3) reported, "The condition approaches the choked disc, there is very great swelling and extreme tortuosity of the veins. The distance between the apex and the base of the disc is represented by a $+\frac{1}{8}$ glass." This referred to only one eye, the other was similar.

On January 6 a very severe attack of pain began. She described it as being at the vertex, and "like anything splitting my head right in two." An injection of morphia gave great relief for three or four hours. At the end of that time she had ten leaches applied, five behind each ear. After this the pain became rapidly more severe.

Temperature 99.4° — 98.4° . Pulse 102—96. Respiration 20—24. For the next four days she suffered the most extreme pain in her head, which made her toss from side to side holding her head in her hands and crying, "Oh, my head! I don't know what I shall do." From time to time she gained relief and even sleep from morphia and chloral. She retched and vomited very frequently. Her temperature varied between 98.4° and 99.4° .

On January 10 her head was actually steaming, vapour rising from it visibly. She had had no application to it. From this time she gradually sank, first into what appeared to be natural sleep, then into heavy sleep, and finally on January 12, into coma, in which she died. The immediate cause of death appeared to be failure of respiration. Her breathing became extremely laboured, her face excessively livid, her hands and

arms also blue and her whole body was bathed in sweat. Pulse excessively weak, 202. Heart's action loud and tumultuous.

Autopsy by Dr. Sutton.—A well developed body. No œdema; fairly nourished. Scalp normal. Very little diploe of skull, bone mostly compact. Dura mater healthy. Sinuses full of blackish blood; even the veins in the bone seemingly distended, and giving the bone over the longitudinal sinuses a deep red appearance. Arachnoid healthy. Pia mater vessels enormously distended and of a blackish colour. It is rare to see the vessels of the pia mater so very much engorged. This was the case mostly over the posterior half of the brain. No wasting of convolutions, nor were they unduly flattened, as is usual in abnormal intra-cranial pressure. On section no disease was appreciable in the brain. Its substance was moderately firm. The grey matter had a pinkish-red, unduly vascular appearance. The central ganglia seemed normal. The pons, medulla, cerebellum, and spinal cord had also in their grey matter the same unduly red hyperæmic appearance. They were firm throughout. No appreciable change in membranes. The optic discs looked swelled, prominent, and their edges lost.

Pleuræ normal. One lung was œdematous. No other change in it. The other was very crepitant, and of a bright red colour. When squeezed only a little blood escaped. On the whole the lungs contained comparatively little blood.

Pericardium normal. Heart normal; left ventricle very firmly contracted, almost empty; right auricle distended with blood.

The other organs were normal, venously congestion only.

The following account of the microscopical appearances of the brain also is by Dr. Sutton.

On examining a section of a convolution of the anterior lobe of the cerebral hemisphere, and contrasting it with a section of healthy brain-convolutions about the corresponding part, it is clear that there is morbid change. It is first noticed that the spherical nuclear bodies in this woman's brain are much more numerous than in the healthy one, whilst the pyramidal cells are very much less numerous. Here and there well formed pyramidal cells are seen with their lateral processes; and immediately adjoining them, where other pyramidal cells

are usually found in a somewhat regular orderly manner, are a large number of well-stained nuclei, about the size of white blood cells, with pale unstained parts (apparently cell-bodies) around them. This paler material is bounded by an ill-defined but unmistakable border. In the border, in some parts, a stellate or spindle shaped nucleus is seen. Although these nuclear bodies have mostly the size just named, some are larger and others much smaller. By the side of some of the larger ones, smaller ones are seen, one or two; as if a larger one had divided, and the smaller were off-shoots. This is a very noticeable feature in many places, and those nuclear bodies seem collected in groups in parts; in other words, they are much more numerous in some parts than in others.

Where more numerous there is very little of the paler substance around them, and around some it is not at all appreciable, the nuclear bodies lying so close as to touch or almost touch each other. Ten or twenty of these may be thus aggregated. These bodies seem to lie mostly in the second and third layers of the convolutions; that is dividing the grey matter of the convolutions into five layers, as is done by Meynezt and others.

The neuroglia as contrasted with that of healthy brain, the sections being about equally thin, is observed to be much more granular; the fibre arrangement is much less distinct, it has a more uniform, granular aspect. The capillaries are for the most part full of red corpuscles. The capillary walls look normal, the perivascular space being very distinct; in parts seemingly wider than normal. There is no appreciable thickening around the larger vessels, and for the most part there is no nuclear growth along the borders of the perivascular spaces, only here and there. There is no decided collections of leucocytes around the capillaries to indicate very active and recent exudation. Here and there, however, two or three round bodies like white blood cells are seen lying on the capillary wall. It is further observed that in some places the neuroglia seems to be disorganised; there are irregularly shaped spaces in which the neuroglia seems to be broken down, whether artificial or otherwise.

Examination of the Backs of the Eyes, by Dr. Gowers.

The backs of the eyes examined after preservation in Müller's fluid.

Eye 1.—The optic nerve entrance is the seat of a swelling of moderate prominence, which extends on each side beyond the edge of the disc, and then gradually subsides to the retinal level. The subsidence is so gradual that it is difficult to measure its diameter; it probably is about the $\frac{1}{10}$ th of an inch. There is a central depression with rather steep sides, the bottom of the depression being $\frac{1}{50}$ th of an inch below the highest portion of the swelling. The greatest thickness of the latter is opposite the edge of the choroid, and there amounts to $\frac{1}{25}$ th of an inch.

The central vein and artery are both large, the vein especially; they show no trace of constriction in passing through the sclerotic ring. They divide close to the bottom of the central pit, the branches approaching close to its surface as they pass up its steep sides, and keeping near the surface over the swelling, but dipping a little at the outer boundary of the swelling.

Section of the swelling shows that it is everywhere infiltrated with small round nucleus-like bodies, $\frac{1}{6000}$ to $\frac{1}{3000}$ inch in diameter, which are aggregated densely in the neighbourhood of the vessels. They are scattered among abundant delicate fibres of connective tissue, which compose the chief bulk of the swelling, and course in various directions. Numerous small vessels exist throughout its substance, and many of the connective-tissue fibres are arranged concentrically around these. Others are distinguishable only by their direction and wavy character from the nerve fibres, which may be traced uninterruptedly through the swelling.

The retina on each side is pushed away from its normal place of commencement, being $\frac{1}{50}$ th of an inch on each side from the edge of the choroidal ring. For a little distance it is greatly thickened, measuring $\frac{1}{40}$ th of an inch in thickness at its commencement. The increased thickness depends chiefly on



Fig. III

F. Burgess, M.D.

W. West & Co., Eng.

Optic Neuritis without Intra cranial Tumour.

thickening of the inner and outer nuclear layers, the inter-nuclear layer being much narrowed. The nuclear layers present only a dense aggregation of such round corpuscles as those already described, and which resemble those normally constituting these layers. The layer of nerve fibres is also thickened, and contains similar nuclei. The retina further outwards gradually assumes perfectly normal characters.

The optic nerve contains many similar corpuscles, some aggregated along the vessels, others lying between the bundles of nerve fibres, others scattered through these bundles. They are more abundant just behind the sclerotic than farther back, but are present in about equal numbers throughout the whole length of optic nerve received.

The section of the bundles shows a granular appearance, the separate nerve fibres being less distinct than usual, but there are no other evidences of degeneration.

The amount of connective tissue between the bundles is not noticeably increased. That in the intervaginal space is however very abundant, the trabeculæ being numerous and thick.

Eye No. 2.—The appearances of this disc, retina, and optic nerve are essentially similar, except that the amount of swelling is rather less (Plate IV, Fig. 3). The height of swelling above the level of the choroidal ring is $\frac{1}{30}$ th of an inch on the side on which the swelling is greatest, and rather less on the opposite side. The commencement of the retina is not displaced outwards so far as in the other eye. There is the same infiltration of the new connective-tissue and of the nerve-fibre layer with minute nuclei.

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